Laboratory tests available for Angioedema Patients

Clinical Presentation		C4 Level	C1-INH Function by Chromogenic Assay	C1-INH Level	C1q Level	C1-INH Autoantibody	Mutation in Factor XII Gene?	C1q Autoantibody	Other tests that may be useful in special cases
Hereditary Angioedema (HAE)		**Inherited defect in the gene for C1-esterase Inhibitor. Autosomal dominant: patients have one normal, one abnormal gene**							
Type I	Recurrent episodic angioedema and abdominal attacks without urticaria. Onset in childhood or young adulthood. 75% have family history. Note that attacks may be estrogen dependent in this group as well as the Type III form.	Usually LOW			NORMAL	NO	NO	NO	C2 LOW, C4a HIGH, C4D:C4 ratio HIGH
Type II		Usually LOW		NORMAL or HIGH					
Type III	Family history present in most cases, onset after childhood. Female patients predominate and attacks often appear to be estrogen-dependent.	NORMAL					YES		Kallikrein or XIIa Function High*
Acquired Angioedema (AAE)		**Condition not inherited but due to other underlying disease. C1-esterase Inhibitor is low due to consumption.**							
Type I	Onset age variable; symptoms same as HAE. No family history, may be associated with underlying lymphoproliferative disease.	Usually LOW		LOW	Usually LOW May be NORMAL	NO	NO		C3 may be LOW
Type II	Onset at variable age; symptoms indistinguishable from HAE. No family history, often associated with an autoantibody that binds to C1-INH.			NORMAL		YES			
Drug-Induced Angioedema		**Angioedema occurs through allergic mechansm and does not involve C1-INH.**							
ACE inhibitors	Pseudoallergic (NSAID) or non-allergic (ACE). Angioedema may start at first use. Acute onset of well-demarcated subcutaneous nonpitting edema of the face or hands.	NORMAL			NORMAL	NO			Trypsin may be HIGH during attack
NSAIDs									
Angioedema with Urticaria		**Angioedema that is accompanied by urticaria. Does not usually involve C1-INH.**							
Allergic AE	Exposure to food, venom, latex, drug, or environmental allergen. May include anaphylaxis.	May be LOW or NORMAL	NORMAL		NORMAL or LOW	NO	NO		Trypsin may be HIGH during attack
AE w/ vasculitis	Longlasting urticaria with tenderness. Vascular damage. With Hypocomplementemic Urticarial Vasculitis the CH50 is usually LOW and so are classical pathway components.						N0, YES with HUVS		CD203c test for chronic urticaria, thyroglobulin or microsomal autoantibodies?
Idiopathic AE	Frequently chronic and relapsing; often occurs with urticaria. Consider test for chronic urticaria (CD203c).				NORMAL		NO		

Most Angioedema (AE) patients can be diagnosed from the clinical presentation supported by these lab tests.

Some patients may require further workup as suggested by these tests.

* Test under development.

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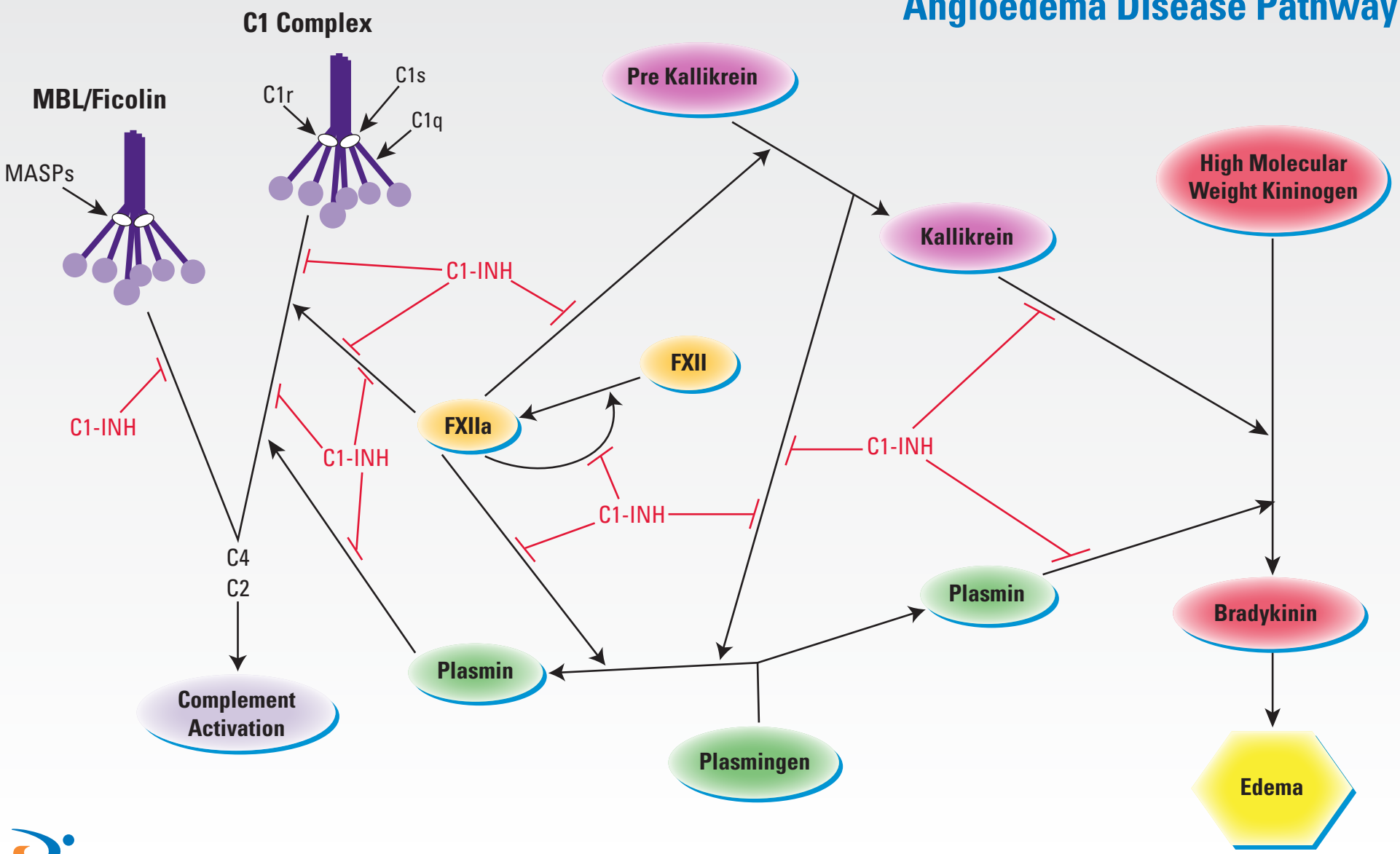
Angioedema Disease Pathway



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